Antibodies to Hair Follicles and Striated Muscle in a Case of Juvenile Dermatomyositis

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Alexander, S., and Stimmler, L. (1971). Archives of Disease in Childhood, 46, 363. Antibodies to hair follicles and striated muscle in a case of juvenile dermatomyositis. A girl of 14 with dermomyositis is described. In addition to the usual muscle and skin lesions she also developed frizzy hair. Her illness was associated with the presence of circulating antibodies to hair follicles and striated muscle.

Dermatomyositis in children is an uncommon disease and one in which autoantibodies have not, to our knowledge, previously been found. We report a case of dermomyositis in a child who showed a qualitative change in her hair, and in whose serum antibody to hair follicle and muscle was found.

**Case History**
This girl was first seen at the age of 10 years 10 months when she complained of difficulty in getting on the bus and in climbing any steep steps. She could walk 3 to 4 hr on the level. Her physical appearance at that time (Fig. 1a) was characteristic of dermomyositis. She had a scaly violaceous rash on both upper eyelids and a faint redness of butterfly distribution on her face. She had capillary dilatation at the upper eyelid margins and the nail beds of her fingers. There was generalized weakness of all muscle groups; the proximal muscles being much more severely affected than the distal ones. Electromyography indicated myositis. Muscle biopsy

![Patient: (a) 10 years and 10 months; (b) at 13 years and 11 months.](http://adc.bmj.com/Archives of Disease in Childhood, 1971, 46, 363.)
showed foci of atrophy accompanied by aggregates of plasma cells and lymphocytes. Rose's DAT test negative. Administration of edrophonium chloride failed to improve muscular power.

A diagnosis of dermatomyositis was made. Because of the pronounced muscle weakness, treatment with prednisone 5 mg t.d.s. was begun. Her skin lesions cleared up and muscle power became completely normal. Unfortunately, she became very obese and cushingoid. Steroid therapy was therefore reduced to a dose of 2.5 mg b.d. which allowed her to climb three flights of stairs at school, at the same time not producing serious side effects. However, on this dose there was still muscular weakness and the skin lesions remained (Fig. 1b). Over the next 2½ years her hair became progressively frizzy (Fig. 1b), having previously been straight. In the past 3 months she has gone into remission with regard to her muscle power. Her hair remains frizzy.

Antibody studies were undertaken 3 years after the start of her illness, and at a time when she was partially treated with prednisolone.

**Methods**

The patient's serum was allowed to react with frozen sections of rat lip (hairless skin), ordinary rat skin (for hair follicles), human group O Rh negative scalp skin, human thyroid tissue, and stripped muscle. After washing with buffered saline, antihuman gamma-globulin labelled with fluorescein isothiocyanate was applied, and after a second washing mounted and examined microscopically by ultraviolet light (Coons and Kaplan, 1950). Sera from normal subjects and from a patient with myasthenia gravis were similarly tested.

**Results**

The results are summarized in the Table.

![FIG. 2.—High power photograph of striped muscle showing positive immunofluorescence of A bands (first serum). (×500).](http://adc.bmj.com/)

Fluorescence of the A bands of striated muscle (Fig. 2) was found with the patient's serum, March 1970 (age 13 yr 10 mth) and that of 2 myasthenia gravis patients.

The patient's serum, obtained 7 months later when she had gone into spontaneous remission as

<table>
<thead>
<tr>
<th></th>
<th>Rat Lip</th>
<th>Rat Skin (sebaceous gland and hair follicle)</th>
<th>Human Scalp</th>
<th>Rat Striped Muscle</th>
<th>Human Thyroid</th>
</tr>
</thead>
<tbody>
<tr>
<td>Present case</td>
<td></td>
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<td></td>
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<td></td>
</tr>
<tr>
<td>At age 13 yr 10 mth</td>
<td>Negative</td>
<td>Positive fluorescence round hair follicle</td>
<td>Positive fluorescence round hair follicle and sebaceous gland</td>
<td>Positive fluorescence of A bands</td>
<td>No antinuclear factor; no thyroid antibody</td>
</tr>
<tr>
<td>At age 14 yr 5 mth</td>
<td>Negative</td>
<td>Negative</td>
<td>Positive</td>
<td>Negative</td>
<td>Negative</td>
</tr>
<tr>
<td>50 normal controls</td>
<td>Negative</td>
<td>Negative</td>
<td>Negative</td>
<td>Negative</td>
<td>No antinuclear factor; no thyroid antibody</td>
</tr>
<tr>
<td>Serum from 3 cases of myasthenia gravis</td>
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<td></td>
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<tr>
<td>Case 1</td>
<td>Negative</td>
<td>Negative</td>
<td>Negative</td>
<td>Positive</td>
<td>No antinuclear factor; no thyroid antibody</td>
</tr>
<tr>
<td>Case 2</td>
<td>Negative</td>
<td>Negative</td>
<td>Negative</td>
<td>Positive</td>
<td>Positive</td>
</tr>
<tr>
<td>Case 3</td>
<td>Negative</td>
<td>Negative</td>
<td>Negative</td>
<td>Positive</td>
<td>Positive</td>
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</tbody>
</table>

*Muscular remission.
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Serum studies in some of these patients have shown the presence of antibodies to the specific tumour and to muscle (Alexander and Forman, 1968; Copeman and Alexander, 1968).

Dermatomyositis in children, however, is not related to malignancy. Previous antibody studies have been negative.

The finding of antibodies to muscle and ‘hair follicle’ in this child with dermatomyositis who suddenly developed ‘frizzy hair’ during the course of her disease seemed significant. It may be that the antibody to hair follicle is just a concomitant of the damaged hair or it may be causally related.

Frizzy hair does occur in other clinical situations. In particular with regrowth of hair after cyclophosphamide alopecia, it is possible that the change from straight to frizzy hair is a sign of hair follicle damage, which in our patient could have been due to anti-hair-follicle antibody. It is also possible that the damaged hair follicle itself might have been responsible for the production of antibody.

Muscle antibodies also occur in patients with myasthenia gravis. This showed no improvement in strength after administration of edrophonium chloride, nor was the muscle histology even remotely like that in myasthenia. Furthermore, corticosteroid therapy does not result in consistent muscular improvement in myasthenia. Again the problem arises whether in either disease the antibody produces the muscle damage or damaged muscle stimulates the synthesis of antibodies.

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REFERENCES